A Rare Tumor on the Abdominal Wall - Dermatofibrosarcoma Protuberans – A Case Report

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Abstract

Dermatofibrosarcoma Protuberans (DFSP) is an uncommon soft tissue tumor typically presenting as a slow growing, firm plaque on the trunk of young adults. It is a rare tumor with an incidence rate of 0.8 to 4.5 cases per million persons per year. It accounts for between 1% and 6% of all soft tissue sarcomas and 18% of all cutaneous soft tissue sarcomas. The majority of DFSPs occur on the trunk (50%), followed by the extremities (35%) and then the head and neck (15%). We are hereby presenting a case report of DFSP which presented as a lesion on the abdomen diagnosed histopathologically and confirmed by immunohistochemistry. A wide local excision was done as a treatment option in this patient. This case is presented here because of the rarity of the entity and the role of histopathology and immunohistochemistry in diagnosing such tumors.

Keywords: Dermatofibrosarcoma protuberans • Histopathology • Immunohistochemistry

Introduction

Dermatofibrosarcoma Protuberans (DFSP) is an uncommon soft tissue tumor that involves the dermis, subcutaneous fat, and in rare cases, muscle and fascia1. It is a rare tumor with an incidence rate of 0.8 to 4.5 cases per million persons per year. It accounts for between 1% and 6% of all soft tissue sarcomas and 18% of all cutaneous soft tissue sarcomas [1]. The majority of DFSPs occur on the trunk (50%), followed by the extremities (35%) and then head and neck (15%) [2]. The tumor typically presents as a slowly growing, firm plaque on the trunk of young adults. It is considered an intermediate-grade malignancy with a low likelihood of metastasis but a high local recurrence rate [3].

Case Report

A 46-year-old female patient presented with history of lesion over the abdomen since 7 years-8 years. Initially started as a small lesion and gradually progressed in size over 6 years-7 years but with sudden increase in size and associated with pain since 1 year. Patient is a known case of hypothyroidism on treatment. No h/o similar complaints in her family. On examination there was a solitary tumour like growth measuring about 5 cm x 4 cm with an erythematous surface with areas of depigmentation within the plaque and verrucous plaques over the surface of the lesion at 2 places with no ulcerations, lesional tenderness or bleeding on examination. No regional lymphadenopathy was noted. A wedge biopsy specimen from the lesion was taken and sent for histopathology with a differential diagnosis of cutaneous tuberculosis *(Lupus vulgaris*), Bowen's disease, Basal cell carcinoma and Malignant melanoma.

As a work-up for tuberculosis, patient was advised chest X-Ray PA view which was normal and mantoux test which showed an induration of 18 mm after 48 hours. CT thorax was normal.

Routine blood investigations (complete blood count, Liver function test, renal function test) and Urine routine were normal.

The histopathological findings showed a tumour characterised by spindle cells arranged in storiform to whorled pattern in the mid-dermis. Individual tumour cells had ovoid to elongated nuclei with moderate to abundant amount of eosinophilic cytoplasm. These findings were suggestive of Dermatofibrosarcoma protuberans.

Based on this report, Immuno histochemistry (IHC) for confirmation of diagnosis was requested. IHC showed tumour cells strongly positive for CD 34. Ki67 proliferative marker labelled about 30% of the tumour cells. These findings were confirmatory of Dermatofibrosarcoma protuberans.

A CT scan of the anterior abdominal wall was done which showed focal irregular thickening in the anterior abdominal wall in the left lateral aspect at the level of the umbilicus which measured 3.0 mm x 8.5 mm (transverse x AP) and a defect noted in the anterior abdominal wall at the level of the umbilicus measuring 1.3 cm with herniation of omentum.

The patient was referred to surgery department and the patient underwent wide local excision and primary closure. Follow-up after 6 months patient showed no clinical evidence of recurrence or metastasis.

Discussion

Dermatofibrosarcoma Protuberans(DFSP) is a mesenchymal tumor which originates from a dermal stem cell or undifferentiated mesenchymal cell with fibroblastic, muscular, and neurologic features. DFSP is a rare tumor with an incidence rate of 0.8 cases to 4.5 cases per million persons per year. It accounts for between 1% and 6% of all soft tissue sarcomas and 18% of all cutaneous soft tissue sarcomas [1]. The majority of DFSPs occur on the trunk (50%), followed by the extremities (35%) and then head and neck (15%) [2]. The regional lymph nodes are rarely involved. The histopathological findings include normal epidermis and in the dermis the tumor cells are arranged in a storiform or intersecting pattern, parallel to the epidermal surface. It is constituted by spindle cells with little pleomorphism and scant cytoplasm. Mitoses are present, but there is no striking mitotic activity. Infiltration to the underlying tissue (subcutis, fascia, muscle, periosteum) is a common feature [4]. An immunohistochemical panel is useful for diagnosing DFSP. CD34 is commonly positive (80%-100% of cases), while FXIIIa, SMA, desmin, S100, and keratins are negative [5].The lung is the most common site of metastasis (4% to 6%) via hematogenous spread [6]. The fibrosarcomatous variant of dermatofibrosarcoma protuberans has a higher risk of local recurrence (14% to 52%) and distant metastases (8% to 29%) [3]. The optimal treatment modality for dermatofibrosarcoma protuberans is Mohs micrographic surgery. Alternatively, dermatofibrosarcoma protuberans can be treated with wide local excision. The chemotherapeutic agent imatinib mesylate is currently FDA-approved for adults with unresectable, recurrent, or metastatic dermatofibrosarcoma protuberans [3].

Conclusion

The present case is being reported here for the rarity of the entity and also to highlight the importance of histopathology and immunohistochemistry in diagnosing the condition. Mohs micrographic surgery is the treatment of choice but in our patient a wide local excision was done as a treatment modality. Follow-up after 6 months patient showed no clinical evidence of recurrence or metastasis.

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